**EXIT Procedure for an Infant with Hypoplastic Left Heart Syndrome with an Intact Atrial Septum**

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**Introduction**

The ex utero intrapartum treatment (EXIT) procedure was first used for patients with tracheal compression from large congenital cervical neck masses.3,4 By partially delivering the fetus while it remains attached to the placenta, the EXIT procedure allows time for resuscitative measures and provides cardiopulmonary support for the infant. While primarily used for cases with congenital airway obstruction, its use has expanded to include infants with congenital heart disease. We describe an EXIT procedure for a neonate with hypoplastic left heart syndrome with intact atrial septum (HLHS/IAS).

**Case**

A 36+5 week GA male with HLHS/IAS was diagnosed prenatally by fetal echocardiography. Earlier attempt at prenatal atrial septostomy at an outside hospital failed secondary to impending premature labor. The treating institution deemed further attempts at fetal intervention too risky for both fetus and mother. Our plan was for an EXIT procedure with intubation and IV access followed by delivery and immediate atrial septostomy in the adjacent OR. The mother underwent general anesthesia for the EXIT procedure in a pediatric hospital. Doppler fetal heart tones were monitored throughout. Sterile technique was maintained for the EXIT procedure. After uterine incision, the fetus was delivered to the clavicles and intubated by a pediatric anesthesiologist with a 3.0 mmcuffed endotracheal tube using a C-MAC 0 blade. After less than 2 minutes on placental bypass, severe maternal hemorrhage and fetal bradycardia of 75 bpm necessitated delivery prior to IV access. The neonate was transported to an adjacent OR where a single dose of endotracheal epinephrine stabilized his vital signs. IV access was obtained, and 15 minutes after cord clamping the neonate underwent sternotomy with catheter-assisted atrial septostomy. Intra-operatively the patient remained stable on low-dose epinephrine and dopamine infusions. Post-operatively, pulmonary hypertension led to hypotension on days 0 and 1 requiring fluid boluses and continued pressor support. By day 7 epinephrine and dopamine infusions were weaned to 0.02 mcg/kg/min and 4 mcg/kg/min respectively. These rates were continued thursday and weekly, but worsening hypotension necessitated colloidal and blood transfusions on days 11 and 12. On day 13 pneumonia was noted, and the patient underwent an exploratory laparotomy with colectomy. The infant deteriorated rapidly post-operatively, and the parents elected for comfort care. At 13 days the infant expired from sepsis secondary to necrotic bowel.

**Discussion**

Prenatal echocardiogram in the second trimester diagnoses 75 to 84% of patients with HLHS. Six to 11% of patients with HLHS also have IAS.3 Survival to hospital discharge now occurs in 79% of patients with HLHS who possess an atrial communication.4 In infants with HLHS/IAS, survival falls to 68% with post-natal intervention, 52% with fetal intervention, and 28% for those who require immediate post-natal intervention.5,6 HLHS/IAS remains uniformly fatal without fetal or neonatal procedures. While fetal intervention shows greater early survival benefits in high risk neonates, it may result in bradycardia or premature birth. Further, it is not always technically possible. The EXIT procedure with immediate neonatal intervention offers an option to decrease the time of hemodynamic instability and theoretically increase neonate survival. With either procedure maternal risks, which include severe hemorrhage and death, deserve particular consideration. Most mothers are healthy with the potential for future offspring. If at any point a procedure threatens maternal life, it should be aborted. In our patient, although initial atrial septostomy successfully created adequate mixing and systemic oxygenation, the patient succumbed to sepsis. This is consistent with the 28% survival rate for patients requiring emergent post natal intervention for HLHS/IAS without the advent of EXIT procedure.6 In these patients death typically results from multi-organ failure, right ventricular dysfunction, sepsis, bleeding, or coronary problems.5,6 More cases would be needed to determine if the EXIT procedure could increase this survival rate. Despite the patient’s death, important lessons can be gleaned; preparedness and continuous multidisciplinary communication is essential for a successful EXIT procedure. Anesthesiologists, surgeons, cardiologists, and neonatologists must be aware of the plan and contingencies. Both operating suites must be prepared in advance. All facilities performing EXIT procedures should have an outline of equipment needed for delivery as well as an airway algorithm. In this case, early onset neonatal bradycardia and maternal instability required emergent delivery prior to IV access; however, open communication between team members allowed for a successful surgery for mother and baby.

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**References**